

Plastic bronchitis mimicking with foreign body bronchus in pediatric patient – A review

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ABSTRACT

Plastic bronchitis (PB) is an extremely rare clinical entity in pediatric age group where an inspissated cast is found in endobronchial airway leading to respiratory distress. It is a large gelatinous or rigid branching airway cast which can lead to life-threatening episodes. The underlying causes for this PB are pulmonary diseases, congenital heart disease (CHD), and a few other conditions. The endobronchial casts are classified into inflammatory and acellular types. Inflammatory casts are most commonly found in primary lung disease, whereas acellular casts are frequently seen in patients with CHD. Removal of this endobronchial cast rapidly improves this condition. Bronchoscopy with bronchial lavage is considered as the gold standard treatment of PB. The patient is managed by rigid bronchoscopy and removal of this cast from bronchus is like foreign body removal from the tracheobronchial tree. Clinicians should have knowledge for correct diagnosis and treatment of PB. This review article discusses about etiopathology, epidemiology, clinical presentations, and diagnosis and treatment of PB among pediatric patients.

Key words: *Bronchial cast, Congenital heart disease, Pediatric patient, Plastic bronchitis, Rigid bronchoscopy*

Plastic bronchitis (PB) is a disease characterized by the formation of the endobronchial casts that fully or partially obstruct the bronchial lumen [1]. It is a rare clinical entity of unknown etiology. It is characterized by the accumulation of fibrinous mucous plugs as a three-dimensional architecture in the bronchial tree. It often partially or completely blocks the airway and leads to respiratory distress [2]. Although the etiopathology of PB is not well understood, pediatric patients with underlying cardiac diseases are prone for developing bronchial casts, particularly after surgical intervention with direct systemic blood flow into the pulmonary circulation [3]. These endobronchial casts are more adhesive than mucous plugs and attain the shape of the tracheobronchial tree [4]. The clinical presentations of the patients are cough, dyspnea, fever, and wheezing which are often confused with inhaled foreign body in the bronchus. Medical treatment for PB includes inhaled or systemic corticosteroids, aerosolized acetylcysteine, aerosolized heparin, and tissue plasminogen which might be used to liquefy and subsequently remove the casts from the tracheobronchial airway. If this fails, bronchoscopic intervention is required for better clearance of the airway.

METHODOLOGY

For searching the published research article, we conducted an electronic survey of the SCOPUS, Medline, and PubMed

databases. The search term in the database included PB in the pediatric patient. The abstracts of the published article were identified by this search method and other articles were identified manually from citations. This manuscript reviews the etiopathology, epidemiology, clinical presentations, and diagnosis and treatment of PB in pediatric patients. This review article presents a baseline from where further prospective trials for PB could be designed and helps as a spur for further research in this rarely encountered clinical entity.

EPIDEMIOLOGY

The prevalence of PB is unknown [5]; however, in the case of Fontan, it is reported between 4% and 14% [6]. Its mortality rate was described among 5–60% cases [7]. PB can affect any age group but is more common among children [8]. Males and females are usually equally affected but Kumar *et al.*, 2018 reported female predominance [9]. The true prevalence of PB is still unknown which might be due to its unusual nature. Until date, there are around 500 cases documented in medical literature [7]. The first known case of PB was described in second century BC by Galen, a Greco-Roman physician, who documented patient that expectorated soft and viscous casts that were thought to be veins and arteries. Fifteen hundred years later, Morgagni identified the expectorated material as mucus casts arising from the endobronchial airway rather than pulmonary veins [10].

To date, the exact incidence is unknown and it is probably an underdiagnosed disease [11]. One study documented that overall mortality rate of PB is 16% and it is up to 29% in patients with cardiac defects. More life-threatening situations are seen in PB with cardiac defects [12].

ETIOPATHOLOGY

PB is an uncommon, potential fatal clinical condition with extensive inspissated mucofibrinous bronchial casts mimicking the three-dimensional architecture of the tracheobronchial airway [8]. Its consistency is sticky and often difficult to remove by bronchoscopic procedure that differentiates this condition from ordinary mucus plug in the tracheobronchial airway [13]. It is characterized by the production of endobronchial casts of protease content, which act as a foreign body along the bronchial tree [14]. It is seen as a complication of respiratory diseases, lymphatic abnormalities, infective cause, and surgery, particularly for congenital heart diseases (CHDs), especially Fontan procedure [1].

PB is a potential fatal pathology which is often seen in patients with a history of Fontan palliative cardiovascular surgery. Fontan surgical technique was designed for pediatric patients having CHDs with single ventricle heart. It establishes passive blood flow through the lungs by diverting both the superior and inferior vena cava circulation directly to the pulmonary arteries and leads to alteration of the pulmonary vascular dynamics. As this Fontan procedure is becoming safer, so the survival rate of the patient is improving with lesser chance of formation of PB in the tracheobronchial tree. PB is seen more frequently, where the formation of casts occludes the bronchial airway without causing stenosis. There is no evidence of reduced bronchial lumen despite several airway cleaning procedures.

Sometimes, Fontan procedure defect improves the pulmonary symptoms but at the same time, there is an alteration of the systemic circulation. Occasionally, the procedure must be reverted, to minimize the fatality of the PB, which is usually a complex situation due to the cardiovascular implications [15]. The raised venous pressure along with dysfunction, loss of integrity in the endobronchial mucosa and increase in the lymphatic vessels pressure of the thorax is usually associated with broncho-lymphatic fistulas with production of the casts in the bronchial tree [16]. Certain conditions associated with PB are cyanotic CHDs, bronchial asthma, lymphatic disorders, pulmonary infections, sickle cell anemia, and lymphoma. Angelos *et al.* studied pediatric PB patients and revealed that 40% of patients had underlying cardiac defects, 31% had asthma or allergic disorders, and 29% had other disorders [12].

The Seear classification of PB is divided into two categories and is useful for the diagnosis and appropriate adjunctive treatment. Type-I PB is described as hypercellular and is usually associated with inflammatory mediators such as fibrin, neutrophils, and eosinophils. Inflammatory conditions of lungs such as asthma, acute chest syndrome, and allergic bronchopulmonary aspergillosis are included in Type-I. Type-II PB is described as a transudative,

mucoid precipitate which increases central venous pressure as in CHDs and lymphatic leakage [17]. Type-II or acellular casts primarily consist of mucin with no acute inflammatory cells and sometimes a few mononuclear cellular components. These casts are usually seen in children with underlying cardiac problems.

Recently, there is another classification of PB on the etiological basis. There are four etiological groups associated with this classification. CHD is associated with acellular mucinous casts. Asthma and atopic disorders are associated with abundant eosinophils, Charcot–Leyden crystals, neutrophils, and fibrin. Lymphatic diseases have chylous casts and sometimes consist of fibrin. Sickle cell anemia is associated with casts which consist of fibrinous material and pigmented histiocytes in the surrounding fluid [15]. Once endobronchial casts are expectorated, it is cohesive and rubbery in consistency than ordinary mucus plug. The cast possesses varying proportion of mucin, fibrin, and cellular materials. The expectorate casts are often mistaken with different food materials such as noodles, chicken, and meat.

CLINICAL PRESENTATION

Patients usually present with mild symptoms of breathing difficulty, cough, and life-threatening conditions like death. The typical clinical presentations of the patient are dyspnea, wheezing, coughing, respiratory distress, and occasionally chest pain or fever [18]. Wheezing is the commonest clinical finding. It is an unusual disease in pediatric age group where cough persists after adequate and prolonged treatment. This failed treatment gives a high suspect of this rare disease. The hallmark for PB is finding casts in bronchial tree during bronchoscopy. Imaging shows atelectasis and infiltrates on affected side of the lungs and often hyperinflation in the contralateral side [19].

The expectorated bronchial cast varies in duration and frequency and sometimes might be complicated with hemoptysis. Some patients are unable to expectorate the bronchial cast spontaneously which might delay or hide the diagnosis. The clinician should intervene early if clinical presentations of airway obstruction are noticed. In severe cases of PB, substantial cast obstruction might lead to fatal outcome such as cardiorespiratory collapse. PB is considered as a differential diagnosis when the patient presents with clinical presentations such as severe airway obstruction, weak breath sounds, persistent, and respiratory distress which cannot be explained by acute respiratory distress syndrome or acute lung injury and the presence of cord-like material in expectorant.

However, the clinical presentations of PB are nonspecific and usually inconsistent with chest X-ray findings. The classification and diagnosis of PB are mainly done on the basis of bronchoscopy and histopathological biopsy of expelled cast. It is an uncommon disease and must be taken into account with recurrent atelectasis and in cases of suspected endobronchial foreign body. Patients with underlying heart diseases are at a greater risk of death whereas with inflammatory process show less severe clinical presentations.

DIAGNOSIS

The radiological study reveals the site of the impaction of bronchial cast and atelectasis or infiltrates. Chest X-ray is often done at initial routine investigation for finding the PB at bronchial airway (Fig. 1). Hyperinflations of lungs are often evident in the contralateral side of lungs. Computed tomography (CT) scan usually shows impacted casts in the bronchial airways. CT scan with axial view of the chest demonstrates the presence of atelectasis with blockage of endobronchial airway in the affected side. The radiological assessment reveals the site for bronchial casts, showing atelectasis, or infiltration, usually in lower lobes. Hyperinflation is usually located on the contralateral side. CT scan allows for visualization of the casts within the tracheobronchial airway.

The diagnosis is usually confirmed by rigid bronchoscopy which demonstrates the obstruction of the endobronchial casts. Endobronchial casts often look whitish-yellowish color and are too thick in consistency to be suctioned out from bronchus. Few patients expectorate the casts spontaneously which is often mistaken for food materials such as noodles or chicken meat [20]. Some casts appear blackish and soft inconsistency. The lung biopsy specimen usually demonstrates mucin cell hyperplasia without any other pathological significance. PB can be seen at all levels of the tracheobronchial tree but is more commonly found in the lower lobe.

Mucoid impaction in the tracheobronchial airway differs from PB in several ways such as mucoid plugs tend to be in the large segmental bronchi of the upper lobe of the lungs, and they are firmly adherent to the bronchial wall and retained rather than being expectorated. Mucoid plug impaction has a strong correlation with bronchial asthma [21]. Imaging showing atelectasis is a differential diagnosis for foreign body aspiration. The exact pathophysiology for PB is not fully understood, but raised pulmonary venous pressure leading to abnormal response of respiratory epithelium which causes excess mucus production might be the cause for PB [22]. As lymphatic leakage is another proposed etiology for endobronchial cast formation, so lymphangiography is considered as a diagnostic tool for patients of Type II PB.

TREATMENT

The treatment of PB is often challenging to the clinician. There has been no intervention proven effective for treatment of PB regardless of the etiology and physical properties of endobronchial casts. Treatment includes removal of casts and addressing the underlying hypersecretory process. Immediate treatment of PB ranges from medical treatment with corticosteroids, different inhaled lytic agents to bronchoscopy, and potentially other surgical procedures. Corticosteroids are often useful in inflammatory casts (Type-I). Bronchoscopic removal of endobronchial casts has been routinely practiced in most of the cases. PB is usually removed by bronchial lavage by bronchoscopic approach which is considered as the standard treatment [23]. The casts found in bronchus are often described as “pudding like” or with toothpaste appearance. Many of

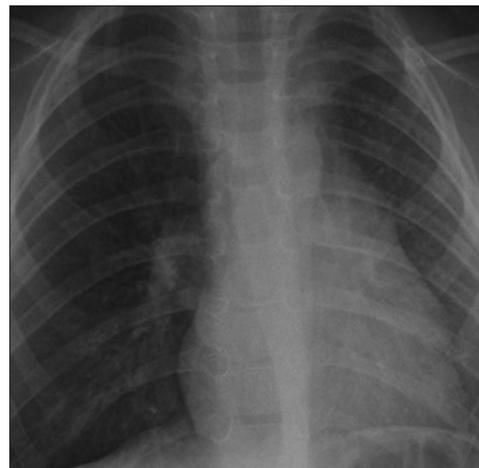


Figure 1: Chest X-ray (Posteroanterior view) showing radiopaque cast in the right endobronchial airway

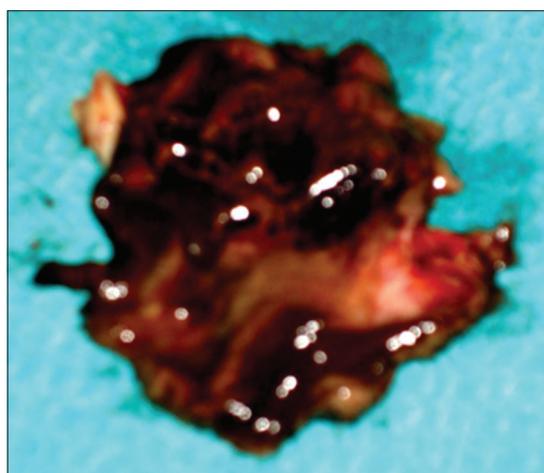


Figure 2: Endobronchial cast after bronchoscopic removal

the casts are too thick to be suctioned out through a bronchoscope or too friable to be grasped and removed by forceps (Fig. 2). Few patients are able to expectorate spontaneously as large casts.

A mucolytic agent such as dornase alfa might be used to liquefy the dense and viscous PB [24]. At present, 7% hypertonic saline solution which attracts water into thick and viscous mucus secretions has good outcome [25]. Chest physiotherapy, acetylcysteine, systemic and inhaled corticosteroids, and DNAase have been used in PB. One recent document showed aerosolized urokinase might be used for treating bronchial casts [26]. Rigid bronchoscopy showed tenacious secretions in the endobronchial airway.

The use of bronchoscopic instruments for the removal of bronchial cast has been documented by Raghuram *et al.* in 1997 [27]. Rigid bronchoscopy has dual role in both diagnosing the casts and giving definitive treatment by suctioning and removing the casts by optical forceps [22]. It gives higher illumination and visualization and maintains the airway by spontaneous ventilation throughout the procedure [28]. Combined suctioning and improved visualization by optical forceps are helpful for complete removal of the endobronchial casts. Although the tenacious consistency and friable nature of the casts is often a

challenge for endoscopic removal, airway clearance is usually done adequately by rigid bronchoscopy.

Bronchoalveolar lavage with rigid suctioning is usually better than optical forceps for friable casts, whereas firmly adherent endobronchial casts are easily removed by optical forceps. This cast can be removed by the bronchoscopy forceps. Premedication with injection atropine helps to reduce airway secretions and prevents from bradycardia in response to airway manipulation by rigid bronchoscope [29,30]. The muscle relaxant is used for controlled ventilation which helps in easier removal of foreign body. Careful intermittent ventilation helpful for proper oxygenation and prevent dislodgment of foreign body or cast further down in the bronchus [31].

Adjunctive treatment beyond rigid bronchoscopy and physiotherapy are usually pharmacological options such as corticosteroids, macrolide antibiotics, inhaled tissue plasminogen activator, and dornase alfa on the basis of the underlying histopathological etiology [32]. In clinical conditions such as lymphatic leakage and CHD and surgical interventions such as thoracic duct ligation and fenestration of the Fontan circuit would prevent cast formation [33]. Recent studies (Benjamin *et al.*) have shown that some patients where underlying comorbidities are not fitting with Seear histopathological classification might present with challenge for finding appropriate adjunctive treatment option [23]. In many cases of PB, rigid bronchoscopy procedure completely clears the endobronchial airway and prevents fatal airway complications.

PROGNOSIS

PB has a poorer prognosis in patients with underlying cardiac defects in comparison to other diseases as the mortality rate is high. Children of PB with allergies or asthma have no life-threatening events. Patients have a greater mortality rate with Type-II (acellular) casts. Deaths in PB with cardiac defects are sudden in nature due to widespread and complete airway obstruction. This makes the care of the children more difficult and needs close follow-up and immediate visit if respiratory symptoms are seen [11]. Overall the prognosis is good in PB except in cases of CHDs where mortality could be as high as 29% and around 41% of the patients present with life-threatening situations as reported by Uchel *et al* [11]. As most of the patients with PB have an underlying disease, it is often difficult to cure cast formation. Majority of the patients require serial bronchoscopic procedure because of complete airway obstruction.

CONCLUSION

PB is an uncommon disease seen in the pediatric age group. If the pediatric patient presents with persistent cough even after adequate and prolonged treatment, a high suspect for this rare disease is mandatory. PB may result in complete obstruction of the tracheobronchial airway which is a medical emergency which needs a multidisciplinary approach for successful diagnosis and treatment. All pediatricians and otolaryngologists should give

special attention for its diagnosis. Endoscopic evidence of PB in the tracheobronchial airway is the gold standard diagnostic procedure. Bronchoscopy along with bronchial lavage is often considered as the treatment of choice.

REFERENCES

- Kruger J, Shpringer C, Picard E, Kerem E. Thoracic air leakage in the presentation of cast bronchitis. *Chest* 2009;136:615-7.
- Walker PA, Shah SK, Letourneau PA, Allison ND, Cox CS. Treatment of plastic bronchitis using serial flexible bronchoscopy and aerosolized heparin therapy. *Eur J Pediatr Surg* 2013;23:157-60.
- Ishman S, Book DT, Conley SF, Kerschner JE. Plastic bronchitis: An unusual bronchoscopic challenge associated with congenital heart disease repair. *Int J Pediatr Otorhinolaryngol* 2003;67:543-8.
- Hasan RA, Black C, Reddy R. Plastic bronchitis in children. *Fetal Pediatr Pathol* 2012;31:87-93.
- Caruthers RL, Kempa M, Loo A, Gulbransen E, Kelly E, Erickson SR, *et al.* Demographic characteristics and estimated prevalence of Fontan-associated plastic bronchitis. *Pediatr Cardiol* 2013;34:256-61.
- Avitabile CM, Goldberg DJ, Dodds K, Dori Y, Ravishankar C, Rychik J, *et al.* A multifaceted approach to the management of plastic bronchitis after cavopulmonary palliation. *Ann Thorac Surg* 2014;98:634-40.
- Eberlein MH, Drummond MB, Haponik EF. Plastic bronchitis: A management challenge. *Am J Med Sci* 2008;335:163-9.
- Brogan TV, Finn LS, Pyskaty DJ Jr., Redding GJ, Ricker D, Inglis A, *et al.* Plastic bronchitis in children: A case series and review of the medical literature. *Pediatr Pulmonol* 2002;34:482-7.
- Kumar A, Jat KR, Srinivas M, Lodha R. Nebulized N-acetylcysteine for management of plastic bronchitis. *Indian Pediatr* 2018;55:701-3.
- Johnson RS, Sita-Lumsden EG. Plastic bronchitis. *Thorax* 1960;15:325-32.
- Uchel T, Labana S, Tancer S, Gonzalez V, Kapadia D, Kulairi Z, *et al.* Unusual case of plastic bronchitis presenting with pneumopericardium. *Am J Case Rep* 2018;19:1434-6.
- Angelos PC, MacArthur CJ. Pediatric plastic bronchitis: A case report and literature review. *Int J Pediatr Otorhinolaryngol Extra* 2010;5:66-9.
- Cajaiba MM, Borralho P, Reyes-Múgica M. The potentially lethal nature of bronchial casts: Plastic bronchitis. *Int J Surg Pathol* 2008;16:230-2.
- Madsen P, Shah SA, Rubin BK. Plastic bronchitis: New insights and a classification scheme. *Paediatr Respir Rev* 2005;6:292-300.
- King G, Ayer J, Celermajer D, Zentner D, Justo R, Disney P, *et al.* Atrioventricular valve failure in Fontan palliation. *J Am Coll Cardiol* 2019;73:810-22.
- Grutter G, Di Carlo D, Gandolfo F, Adorisio R, Alfieri S, Michielon G, *et al.* Plastic bronchitis after extracardiac Fontan operation. *Ann Thorac Surg* 2012;94:860-4.
- Somers E, Day W, Derkay CS. Plastic bronchitis resulting in complete bronchial obstruction in an otherwise healthy child. *Int J Pediatr Otorhinolaryngol Extra* 2017;18:1-4.
- Ugaki S, Lord DJ, Sherwood MC, Winlaw DS. Lymphangiography is a diagnostic and therapeutic intervention for patients with plastic bronchitis after the Fontan operation. *J Thorac Cardiovasc Surg* 2016;152:e47-9.
- Barber BJ, Burch GH, Tripple D, Balaji S. Resolution of plastic bronchitis with atrial pacing in a patient with Fontan physiology. *Pediatr Cardiol* 2004;25:73-6.
- Bowen A, Oudjhane K, Odagiri K, Liston SL, Cumming WA, Oh KS, *et al.* Plastic bronchitis: Large, branching, mucoid bronchial casts in children. *AJR Am J Roentgenol* 1985;144:371-5.
- Benjamin JL, Rychik J, Johnstone JA, Nadolski GJ, Itkin M. Cost-effectiveness of percutaneous lymphatic embolization for management of plastic bronchitis. *World J Pediatr Congenit Heart Surg* 2019;10:407-13.
- Silva RC, Simons JP, Chi DH, Yellon RF, Alper CM. Endoscopic treatment of plastic bronchitis. *Arch Otolaryngol Head Neck Surg* 2011;137:401-3.
- Grizales CL, González LM, Castrillon MA, Sua LF, Lores J, Aguirre M, *et al.* Plastic bronchitis: A case report. *Respir Med Case Rep* 2019;28:100876.
- Jones AP, Wallis C. Dornase alfa for cystic fibrosis. *Cochrane Database Syst Rev* 2010;3:CD001127.
- Donaldson SH, Bennett WD, Zeman KL, Knowles MR, Tarran R, Boucher RC, *et al.* Mucus clearance and lung function in cystic fibrosis with hypertonic saline. *N Engl J Med* 2006;354:241-50.

26. Quasney MW, Orman K, Thompson J, Ring JC, Salim M, Schoumacher RA, *et al.* Plastic bronchitis occurring late after the Fontan procedure: Treatment with aerosolized urokinase. *Crit Care Med* 2000;28:2107-11.
27. Raghuram N, Pettignano R, Gal AA, Harsch A, Adamkiewicz TV. Plastic bronchitis: An unusual complication associated with sickle cell disease and the acute chest syndrome. *Pediatrics* 1997;100:139-42.
28. Swain SK, Panigrahi R, Mishra S, Sundaray C, Sahu MC. An unusual long standing tracheal foreign body a rare incidence. *Egypt J Ear Nose Throat Allied Sci* 2015;16:91-3.
29. Swain SK, Sahoo S, Sahu MC. From tooth extraction to fatal airway complication in a child a case report. *Egypt J Ear Nose Throat Allied Sci* 2016;17:27-9.
30. Swain SK, Mohanty S, Behera IC, Sahu MC. Safe and effective anesthesia during pediatric rigid bronchoscopy: An experience at a tertiary care center of Eastern India. *Pediatr Polska* 2015;90:470-3.
31. Swain SK, Mishra S, Dash M. An unusual asymptomatic foreign body at tracheobronchial tree. *Int J Phonosurglaryngol* 2014;4:40-2.
32. Jasinovic T, Kozak FK, Moxham JP, Chilvers M, Wensley D, Seear M, *et al.* Casting a look at pediatric plastic bronchitis. *Int J Pediatr Otorhinolaryngol* 2015;79:1658-61.
33. Wilson J, Russell J, Williams W, Benson L. Fenestration of the Fontan circuit as treatment for plastic bronchitis. *Pediatr Cardiol* 2005;26:717-9.

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